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REMARKS
ON
CYSTIC LYMPHANGIOMA IN CHILDHOOD:
ITS
DIFFERENTIAL DIAGNOSIS AND TREATMENT.[1]

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CYSTIC lymphangiomata may occur in any part of the body, and they are not unusual in the neighbourhood of the axilla. They are least frequent on the face, but they are seen as congenital or acquired tumours in the tongue, where they cause macroglossia, and even in the mucous membrane of the intestines. They present many points of interest wherever they occur. Their pathology, their mode of growth, and their anatomical peculiarities are all subjects which might be considered with profit, but I shall restrict myself to the points of clinical interest; and it seems to me that the two great points which have to be answered about such tumours are: How are they to be recognised? And, when they have been recognised, How are they to be treated?

A cystic lymphangioma varies from a few scattered vesicles growing from the skin and filled with lymph to a tumour like the one you see. The scattered vesicles to which the fanciful name of "lupus lymphaticus," and the more correct one of lymphangioma cutis circumscriptum, has been given, is not likely to be mistaken for anything else, but the more solid forms of tumour must be distinguished from enlarged lymphatic glands, from a fatty tumour, from a nævus or other form of blood cyst, from a dermoid and from a sarcoma, for any of these swellings may be found in a newly-born child. They all increase in size shortly after birth, and, in the early stage at any rate, none of them are necessarily associated with any marked alteration of the skin covering them. Each of these tumours, too, may require surgical treatment.

Local hypertrophy of a part which is known as macrodactyly, either general or due to the overgrowth of a single tissue is perhaps most likely to be mistaken for a cystic lymphangioma occurring in one of the extremities. Such a case was under the care of Mr. Robert Jones, of Liverpool, who recently asked me to bring it before one of the London

[1] Read at a Meeting of the Metropolitan Counties Branch, North London District.

societies. The patient, a girl, aged 18 months, presented a tense and elastic swelling, covered by healthy skin, on the hand. It invaded the thumb, the forefinger, and the radial side of the right hand, rendering the thumb and finger so useless that they had to be amputated. The swelling was smooth and uniform, and its growth was so slow and steady that its nature was correctly diagnosed before the operation was undertaken. Subsequent microscopical examination showed that the whole of the subcutaneous connective tissue, and even the intermuscular planes were infiltrated with adipose tissue, which did not differ in any respect from healthy fat. The bones are thin and elongated, but the change is, I think, secondary to the increased deposit of fat, and did not imply that they were involved in the hypertrophy. The enlargement, therefore, was due to an overgrowth of fat which for some obscure reason had increased out of all due proportion over this very limited area of the body. Other diffuse lipomata also occur in children. They are deeply placed amongst the muscles of the limb, and they grow from the outer layers of the periosteum.

These forms of diffuse lipomata are much more likely to be mistaken for cystic lymphangiomata than are the lobulated and well circumscribed swellings, which there is no difficulty in recognising, for they present the characters of an ordinary fatty tumour. Cystic lymphangioma may be distinguished from diffuse lipomata by the greater tenseness of the lymphatic swelling and by the peculiar elastic sensation which is given when a number of cysts are felt beneath the skin, whilst the lipomata are softer and more doughy. The skin too is more intimately blended with the lymphatic tumour than with the lipoma unless the cyst lies beneath the deep fascia, when the skin is freely movable and is absolutely unaffected. Lymphangiomata usually present prolongations of their tissue, which may run considerable distances either deeply or superficially, and they are thus less well defined than even diffused fatty tumours. These prolongations are sometimes represented by isolated tumours, which are separated from the main growth by an area of healthy tissue, as was the case in the girl you have seen to-day. The recurrent attacks of lymphangitis, if they are present, will go far to prove the lymphatic origin of such a tumour, but the surgeon must be on his guard lest he mistake these attacks for true erysipelas.

Cystic lymphangiomata have also to be distinguished from cavernous nævi and those rarer forms of blood tumours which arise either subcutaneously or much more deeply beneath the muscles as pockets or pouches in the veins, like the botryoidal appendages seen in some of the vermes. Cavernous nævi very closely resemble the cystic lymphangiomata, from which indeed they only differ in being developed from the vascular instead of from the lymphatic system. Nævi, however, are more common than lymphangiomata, they can always be emptied, or at any rate they can be reduced in size by well regulated pressure applied for some length of time, whilst muscular exertion and all efforts leading to venous engorgement of necessity increase their size. Some nævi have a distinct varicocele-like feel, and in some the skin is the seat of a capillary nævus or it presents other evidence of disturbance of the vascular system. Nævi in which there is much connective tissue and those rarer forms

containing fatty or sarcomatous tissue are less easy to diagnose from cystic lymphangiomata, and in such cases an exploratory puncture will alone give a correct diagnosis. The lymphatic changes sometimes go hand in hand with the vascular ones, so that the cystic lymphangioma is at the same time a cavernous nævus, whilst in other cases the lymphatic cysts may contain a blood-stained fluid. The relation of lymphangiomata to "degenerate nævi" is an interesting one, but it need not detain us to-day.

Dermoid tumours are frequently met with in children, but they must not be mistaken for lymphangiomata even though they contain loculated cysts. The walls of a dermoid are thick, and they have a much more resistant contents than the serous fluid filling lymphatic cysts. Some dermoids are more likely to be mistaken for the extensive lymphatic tumours to which allusion has already been made under the name of cystic hydrocele of the neck. Indeed, in the earlier stages, before the skin has become thinned, it may be impossible to distinguish with any certainty between the two conditions. Dermoids, however, are always situated where the developmental processes are more complex than usual; they are more regular in shape, they are better circumscribed, they grow more slowly and more uniformly than lymphatic tumours.

Sarcomata are not unusual in children; they may be congenital, but much more often they occur some weeks after birth; they increase rapidly in size, the skin is soon involved, they fungate, and dissemination is a very early feature. These characters are usually so well marked that, except in the earlier stages, a sarcoma should not be mistaken for a cystic lymphangioma. The difficulty of diagnosis may be great, but the more rapid growth of the malignant tumour would be an important help in distinguishing between the two conditions.

History of a Case.

N. W., then a fortnight old, was admitted into the Victoria Hospital for Children on August 16th, 1894. The photograph, (Fig. 1) taken on the day after her admission, shows that she had a large swelling which involved the whole of the left side of the face and the upper half of the neck. It was painless, soft, and roughly lobulated. There was an isolated tumour with the same characters behind and just below the lobule of the ear. It was as large as a walnut, and seemed to be cystic. The skin was stretched over both the tumours, and was natural in colour and in consistence. It did not present any warty growth, but it was adherent to the swellings, which could be moved upon the deeper structures. There was no œdema of the tissues, nor were the veins enlarged. The mucous membrane lining the cheek and lying immediately beneath the swelling was healthy. The neighbouring lymphatic glands were unaffected.

The mother said that the left cheek was larger than the right when the child was born, but that both cheeks then felt equally soft. The left side of the face increased rapidly in size, and the swelling became harder as it got larger. The child took the breast well, and had thriven. An elder child was healthy in every respect, and there was no history of any similar tumour in the family. The mother was very delicate, and she had been ailing during the whole of her pregnancy.

The patient was put under the influence of chloroform on August 20th, and incisions were made over the tumour so as to remove an elliptical piece of skin measuring about 2 inches in its widest diameter. The long axis of the ellipse was vertical and extended from top to bottom of the large tumour. As soon as the skin was divided the tumour was seen to consist of numerous cysts varying in size from a pea to a pigeon's egg, each containing a clear serous fluid unmixed with blood. The cysts were separated by septa of fibrous tissue, but there was no definite capsule. It was continuous with

Fig. 1.—Photograph of a child, aged 14 days, with a cystic lymphangioma of the cheek.

the deeper layers of the skin, and involved the connective tissue beneath it. As it was impossible to dissect out the whole tumour, the cysts were removed as completely as possible, and the gaping walls of the remainder were cut away. Many filaments of the facial nerve were involved in the tumour, and some of them had to be divided; the larger branches, however, were carefully avoided, and those going to the orbicularis palpebrarum were recognised and preserved. The tumour was thoroughly removed from the cheek, but a portion was left upon the temple, and the iso-

lated tumour behind the ear was not touched. The skin was drawn together with interrupted sutures of horsehair. The two edges of the wound came together accurately on the face, but there was a considerable excess of skin in the neck, so that it was thrown into folds. I did not regret this, for the skin of children retracts readily, and Nature is very kind in rendering their rough places smooth. A tube was put into the lower end of the wound, and a dressing of cyanide gauze was applied. The child was somewhat collapsed after the operation as it lost a considerable amount of blood in spite

Fig. 2.—Photograph of same child seventeen months after removal of the tumour.

of the prompt application of pressure forceps to the bleeding points, but it soon recovered. The tube was removed when the wound was dressed two days later, but no further dressing was required until August 31st, more than a week afterwards. The wound had then healed so completely that the sutures were removed, and the child was sent home.

The child was again admitted to the hospital two months later for an attack of diarrhœa, from which she recovered under appropriate treatment.

She was readmitted to the hospital on June 26th, 1895, when her mother stated that for some weeks previously she

had had recurrent attacks of inflammation of the face and neck upon the affected side. The skin became red and eczematous, and a clear fluid exuded from the inflamed surface, forming crusts, whilst the surrounding tissues became swollen. The attacks recurred every ten days or a fortnight, and lasted for three or four days. They were not accompanied by any feverish symptoms. The child had one of these attacks whilst she was in the hospital, and we thus had an opportunity of confirming the accuracy of the mother's account and of ascertaining that there was no rise of the body temperature. Such recurrent attacks of lymphangitis are said to be characteristic of cystic lymphangioma, and it is also said that the tumours then grow rapidly, but I could not satisfy myself that the swelling behind the ear or that part which remained upon the temple had sensibly increased in size since I had last seen the patient.

On July 15th, 1895, I removed the tumour behind the ear, and with it some of the redundant skin left after the first operation. The wound was dressed on July 17th; it had healed by first intention and without any rise of temperature. The stitches were removed, and a few days afterwards the child was sent home cured. Fig. 2 is a photograph of the child taken in December, or seventeen months after the last operation. It shows that there is still some fulness over the left temple.

I examined microscopical sections of the tumour after a portion of it had been hardened in Foà's solution and in alcohol. The epithelium was normal in every respect, but even the connective tissue forming the papillæ of the true skin contained dilated lymphatic spaces, so that it was easy to see how a further slight dilatation might give rise to the vesicles often seen upon the skin in these cases. The deeper layers of the cutis vera surrounding the sweat glands were riddled with large lymphatic sinuses. Each sinus contained unicellular lymphoid cells with coagulated lymph, whilst in some cases a few red blood corpuscles were present, but they appeared to have gained admission accidentally. The walls of the sinuses consisted of a layer of endothelial cells, external to which was a coat of connective tissue. This coat contained in nearly every instance a larger or smaller number of unstriped muscle fibres. The sinuses were usually well circumscribed, but in a few instances the walls were deficient at some part of their circumference, so that the sinus was in direct connection with the lymphatic spaces in the surrounding connective tissue. There was a good vascular supply to the tumour, and the arteries were remarkable for the thickness of their walls, a thickness due partly to an increase in the connective tissue elements of the middle coat and partly to an increase in the subendothelial layer. The sweat glands, the hairs with their follicles, and the adipose tissue appeared to be normal in every respect. There was no formation of lymphadenoid tissue in any part, nor, if the repeated inflammation to which the tumour had been subjected be taken into consideration, was there an undue proportion of small round cells in the connective tissue of the growth. The tumour was, therefore, a cystic lymphangioma agreeing in most of its histological characters with that described by Mr. Harold Stiles in the first volume of the Edinburgh *Hospital Reports*. The absence of a part of the wall in some of the cysts is interesting, for it shows how the more rare form of

cystic lymphangioma may pass directly into a cystic hygroma, which is a little more common. The lymphatic swelling in a cystic hygroma or hydrocele of the neck consists of a single cyst, either simple or lobulated, which is deeper in its origin and has more complex relations than a cystic lymphangioma.

The treatment of cystic lymphangiomata, like the treatment of most surgical conditions, is open to the widest divergence of opinion. It appears that some cysts, even when they have attained to a considerable size, may disappear spontaneously, but it is unreasonable to expect such a disappearance in any individual case; and I believe that the proper course to adopt in this child was that of excision of the tumour. It seemed at the time a severe measure, for the swelling was situated upon the cheek, where of necessity nothing could be done to arrest hæmorrhage except the prompt application of pressure forceps to the bleeding points. The child, too, was very young; on the other hand it was healthy, the swelling was very unsightly, and the parents were anxious that an operation should be performed if it offered a reasonable chance of success. The result more than realised my expectations, for the wound healed by first intention, though it often happens that union is not kindly in such cases. As in all operations upon young children, where there is likely to be much loss of blood, an attempt was made to diminish the shock by putting the patient during the operation upon a water bed half filled with water at 110° F., and afterwards by keeping her more than usually warm.

The position of the tumour and the frequent attacks of lymphangitis prevented me from using, in place of the second operation, any of the methods which are available for the treatment of the larger nævi. Electrolysis at its best would have produced a fibrous tumour, and, this being removed, would have left a scar no less marked than that resulting from excision of the original cyst. Setons, even of the finest thread, I look upon as relics of a bygone surgery, whilst puncture and drainage I felt sure would be ineffectual. Simple puncture may be effectual in cases of hydrocele of the neck, for the tumour is only a single cyst, and in such cases it should always be tried before any more formidable operation is decided upon. It is easy to keep the cyst aseptic after such a puncture, and, if the fluid again collects, the patient is in no worse position than he was before, whilst if, as I have seen, the cyst does not refill the child is saved from an operation which always puts it in peril of its life either during the operation or from subsequent complications.